

Approach to Nutcracker Syndrome

Nutcracker syndrome

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Nutcracker syndrome (left renal vein entrapment) is a rare disease where there is symptomatic compression of the left renal vein (LRV) between the aorta and the superior mesenteric artery. LRV entrapment divided into 2 types: anterior and posterior. Typical nutcracker morphologic features imply compression of the left renal vein (LRV) between the abdominal aorta and the superior mesenteric artery (SMA) known as anterior nutcracker, whereas compression between the aorta and the vertebral column refers to posterior nutcracker syndrome [1-3].

Nutcracker syndrome (NCS) is a rare disease and because of the variety of symptoms, the true prevalence remains unknown and it may be underdiagnosed [2,3]. Although most symptomatic patients are women in their third or fourth decades of life, the syndrome is also described in adolescents and men. Clinical presentation vary from asymptomatic microhematuria to severe pelvic congestion [2]. Patients usually present with nephrological or urological symptoms. Microscopic or macroscopic hematuria, orthostatic proteinuria and left side pain are common symptoms of nephrologic presentation. Abdominal pain, varicocele, dyspareunia, dysmenorrhea, orthostatic intolerance and fatigue are the components of the urologic presentation [2,3].

The primary diagnostic step must be a careful physical examination and suspicion of the syndrome by excluding other causes of abdominal pain. Doppler ultrasonography with color flow is the first preferred non invasive diagnostic test for NCS. Doppler ultrasonography shows the course and the blood flow velocity of the LRV [3]. The peak velocity ratios of the aortomesenteric and hilar portions of the LRV higher than 4.2 is one of the diagnostic criteria [3,4]. Contrast-enhanced computed tomography (CCT) or magnetic resonance imaging (MRI) provides additional information. These imaging tools provide visualization of the anatomy, course of the LRV and can demonstrate the LRV compression point. The ratio between the diameters of the aortomesenteric and hilar portions of the LRV (beak sign imagination) above 4.9 is considered significant. In addition, the aortomesenteric angle is about 90 degrees in the normal population [3,4]. In NCS, this angle will be narrower. Aortomesenteric angle less than 35 degrees is a highly suggestive sign of NCS [3,4]. Retrograde transcatheter angiography is the first choice among invazive diagnostic methods. In healthy individuals there is no significant pressure gradient across the LRV. It is usually higher than 1 mmHg in NCS [2-4].

The treatment options of NRS are in a wide spectrum and are ranged from conservative follow up to nephrectomy. Management depends on the severity of symptoms and compression. Mild abdominal pain does not interfere with short daily activities can be treated conservatively despite microscopic hematuria and orthostatic proteinuria. In the same way patients less than 18 years of age are managed conservatively. It is believed that increased retroperitoneal fat and fibrous tissue leads to anterior displacement of the kidneys and further increase the aortomesenteric angle in adolescents [3,4]. Until such relief is met, analgesics may be used for pain control, angiotensin converting enzyme inhibitors have been tried to improve proteinuria and aspirin therapy to prevent venous thromboemboly.

LRV stenting as interventional treatment may be preferred to open surgery. However compression of the stent between AA and SMA, potential complications of stenting such as thrombosis susceptibility, migration, fistulization, need for long term anticoagulant therapy and endothelial fibrous hyperplasia limits its use; besides long term follow-up results are lacking [4].

A variety of surgical techniques have been proposed for NCS, including LRV transposition, nephropexy, renal autotransplantation and gonadal vein bypass [2-4]. The surgical treatment protocol can be designed according to the anatomical structure of the patient and the experience of the clinic. In transposition technique, the aim is to transport LRV distally into the IVC and relieve the compression of LRV between AA and SMA. Renal auto-transplantation involves nephrectomy and transplantation of the kidney into either ipsilateral or contralateral iliac fossa. Anterior nephropexy with excision of renal varicosities and gonado-caval bypass has been performed in patients with NCS associated with pelvic varices but has not gained wide popularity. In addition, simple nephropexy with excision of varicosities is no more recommended as it fails to address the primary pathology. Surgical techniques described for renal vein should occasionally include interventions such as ablation of pelvic venous collaterals or coil embolization of ovarian veins in patients with pelvic congestion and genital varices.

A very high risk exists for misdiagnosing as urinary tract infections, nephropathy, idiopathic hypercalciuria and urolithiasis, resulting in unnecessary medical treatment and surgery. Diagnosis of NCS is challenging, but an appropriate treatment can spell relief.

Nutcracker syndrome is a rare disease and clinical suspicion constitutes a basis for the diagnosis. For the the definite diagnosis, other more common causes of abdominal pain must be ruled out. Management of NCS depends upon the clinical presentation and severity of symptoms. Decision of surgical treatment and choice of the technique should be made meticulously for every patient individually.

References

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